

## CHAPTER 17

# THE WOMAN WITH DAILY PERSISTENT HEADACHES

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## Case History

A previously healthy and headache-free 30-year-old woman developed bronchitis. During a coughing spell she suddenly developed a severe holocephalic headache. Because of the severe pain she came to the emergency department, where she had a normal neurologic examination. Routine laboratory tests were normal, as were a computed tomography (CT) scan and lumbar puncture (LP). The patient then developed a severe lumbar puncture headache and was hospitalized. Magnetic resonance imaging (MRI) of her brain was normal, and an otolaryngology consultation failed to identify any abnormality. The positional component of the headache was successfully treated with intravenous caffeine, but there was no other clinical improvement during her hospitalization.

Several weeks later, the patient went to a headache center. By this time, the severity of her baseline daily, holocephalic headache was moderate with exacerbations associated with nausea and photosensitivity. Brief as well as prolonged exacerbations of pain were triggered by coughing, sneezing, and Valsalva's maneuver. She was unable to work.

## Questions about This Case

- What is the diagnosis and the differential diagnosis?
- What other tests would you do?
- What treatments would you try?

## Case Discussion

A 30-year-old woman developed chronic daily headache without any prior history of headache. The sudden onset prompted the appropriate evaluation for subarachnoid

hemorrhage, meningitis, intracranial mass lesion, and most systemic illnesses.

The onset of a daily headache without a background of worsening episodic headaches should prompt a thorough investigation for secondary causes of headache, even with normal general and neurologic examinations. An extensive list of secondary causes of new daily persistent headache (NDPH) can be generated.

Most other diagnostic possibilities were excluded in this patient by the tests performed. We ordered a magnetic resonance venogram to exclude venous sinus thrombosis and a Lyme titer—both were negative. A diagnosis of NDPH was made.

Our patient's acute onset of headache with cough is unusual but is consistent with the proposed criteria for NDPH, which require onset over 3 days or less.

Because of our patient's cough headache, indomethacin was tried but it was unsuccessful. The patient was hospitalized and treated with the repetitive intravenous dihydroergotamine protocol and started on methylergonovine as a headache preventive. She became headache free in the hospital and returned in follow-up with a biweekly episodic headache. She has been weaned off her preventives and continues to have moderately severe episodic headache, but is now back at work.

## Management Strategies

- Establish the correct diagnosis.
- An MRI and a Lyme titer are indicated in this case to rule out secondary headaches.
- Avoid analgesic overuse—rebound may occur in NDPH.
- Hospitalization is indicated if the patient is disabled and has been refractory to aggressive outpatient strategies.

- Always bear in mind that a secondary headache might exist—with this pattern of headache onset an extra level of vigilance is indicated.
- Treat with abortive and preventive agents that work for transformed migraine or chronic tension-type headache.

### Case Summary

- The patient has NDPH.
- In this case, as with many NDPH cases, onset occurs with a systemic illness, however, the nature and treatment of this systemic illness appear to have little bearing on the treatment and final outcome of the daily headache which persists long after the original illness is gone.
- There is little literature to help the clinician provide the most successful abortive and preventive strategies.
- Inpatient treatment with repeated intravenous dihydroergotamine is often successful.

### Overview of New Daily Persistent Headache

Daily headache may begin without a history of evolution from episodic headache. Based upon the absence of identifiable structural or metabolic causes for headache in many of these patients, Vanast proposed an entity that he called new daily persistent headache (NDPH). Silberstein et al. found that NDPH accounts for a significant number of clinic and hospitalized headache patients. NDPH has been found in children and adolescents as well as adults. Silberstein et al. have proposed that NDPH be added to the International Headache Society taxonomy as a separate primary headache disorder and put forward the criteria listed in Appendix 17–1.

Note that these criteria, like those of the other primary headache disorders, require the exclusion of secondary causes of headache (Appendix 17–2).

### Selected Readings

- Gladstein J, Holden EW. Chronic daily headache in children and adolescents: a 2-year prospective study. *Headache* 1996; 36:349–51.
- Hamada T, Ohshima K, Ide Y, et al. A case of new daily persistent headache with elevated antibodies to Epstein-Barr virus. *Jpn J Med* 1991;30:161–3.
- Mathew NT. Chronic refractory headache. *Neurology* 1993;43 (6 Suppl 3):S26–33.
- Silberstein SD, Lipton RB, Sliwinski M. Classification of daily and near-daily headaches: field trial of revised IHS criteria. *Neurology* 1996;47:871–5.

Silberstein SD, Lipton RB, Solomon S, Mathew NT. Classification of daily and near-daily headaches: proposed revisions to the IHS criteria. *Headache* 1994;34:1–7.

Vanast WJ. New daily persistent headaches: definition of a benign syndrome. *Headache* 1986;26:317.

### Editorial Comments

*Sometimes patients develop new headaches out of the blue without a previous history of migraine or other primary headache disorders. These headaches can persist on a daily basis and, in the absence of any specific etiology, have been termed new daily persistent headache or NDPH. Dr. Young presents such an intriguing and interesting case and suggests the diagnosis of NDPH. It is still not certain in our minds that this patient does not have a secondary cause for her headache, but we are in full agreement with the approach taken and the overall philosophy regarding diagnosis, put forward by Dr. Young. Time will be the ultimate judge of whether cases such as this are NDPH or some other disorder that we cannot detect at present.*

### Appendix 17–1: Criteria for New Daily Persistent Headache, Proposed by Silberstein et al.

#### 4.7 New Daily Persistent Headache (NDPH)\*

- Average headache frequency 15 days/month for >1 month
- Average headache duration >4 hours/day (if untreated). Frequently constant without medication but may fluctuate.
- No history of tension-type headache or migraine that increases in frequency and decreases in severity in association with the onset of NDPH (over 3 months)
- Acute onset (developing over <3 days) of constant unremitting headache
- Headache is constant in location? (Needs to be tested)
- Does not meet criteria for hemicrania continua (4.8)
- At least one of the following:
  - There is no suggestion of one of the disorders listed in groups 5–11 (this refers to IHS diagnostic groups)
  - Such a disorder is suggested, but it is ruled out by appropriate investigations
  - Such a disorder is present, but the first headache attacks do not occur in close temporal relation to the disorder

\* May occur with or without analgesic overuse (rebound)

## Appendix 17–2: Differential Diagnosis of NDPH

1. Intracranial structural
  - Tumor
  - Stroke—particularly hemorrhagic
  - Epidural, subdural hematoma
  - Venous sinus thrombosis
  - Post-traumatic headache
2. Disease of skull, spine, temporomandibular joints
3. Sinusitis, especially sphenoid sinusitis
4. High and low pressure headaches
5. CNS infection
  - Lyme disease
  - Atypical bacterial/viral (shunt) infection
  - Fungal meningitis
6. Chemical meningitis
  - Noninfectious nonsteroidal-drug-induced meningitis
7. Hypercarbia/obstructive sleep apnea
8. Systemic illness
  - Infections: EBV, Lyme, HIV
  - Autoimmune disease

- Temporal arteritis
  - Carcinoid syndrome
9. Toxic metabolic
    - Medication-induced
    - Intoxication, especially carbon monoxide
  10. Psychogenic

The clinical features, other than daily headache duration, are not specified in Appendix 1. In our experience, NDPH often resembles chronic tension-type headache or transformed migraine. It may develop during the acute or recovery phase of a flu-like illness.

Several investigators have searched for the causes of or trigger factors for NDPH and have suggested that particular viruses may be involved in its genesis. I do not routinely look for these abnormalities since they will not alter therapy.

Unfortunately, little has been written about therapy for NDPH. Many clinicians have the impression that NDPH is quite difficult to treat and less responsive to therapy than correspondingly severe transformed migraine. Several investigators have noted that this headache responds to inpatient repetitive intravenous dihydroergotamine. For the most part, clinicians tend to treat this disorder similarly to chronic tension-type headache or transformed migraine.